Case Report

Adult Type Anomalous Left Coronary Artery from Pulmonary Artery

GP Parale*, SS Pawar**

Abstract
Anomalous origin of Left Coronary Artery from Pulmonary Artery (ALCAPA) is a rare congenital anomaly. Its survival into adulthood is further rare. Clinical manifestations result from evolving morphological-functional alterations in pulmonary circulation that occur after the birth. We report a case of a 43 year old adult patient with effort angina and without any ECG or Echo abnormalities. On coronary angiography, typical anatomy of ALCAPA was revealed. ©

INTRODUCTION
Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) is a rare congenital defect with reported incidence of 1 in 3 lac live births accounting for 0.25% of congenital heart diseases1 and survival to adulthood is furthermore rare accounting for only 10-15% of these cases.2 Here, we report a case of adult survivor of ALCAPA diagnosed at our institution.

CASE REPORT
A 43 year old male hypertensive since two years and known chronic smoker was evaluated for history of class II effort angina over previous two months. Physical examination was within normal limits except for blood pressure, which was 156/108 mm of Hg in right upper arm in supine position. X-ray chest was not remarkable. The ECG showed ST/T changes in leads I, avL and V6 suggesting lateral ischaemia. 2-dimensional echocardiography revealed trivial mitral regurgitation (MR) and concentric left ventricular hypertrophy (LVH) with preserved left ventricular systolic and diastolic function. There was no regional wall motion abnormality. [Echocardiography was done before as well as after the angiography with prior knowledge of coronary artery anatomy. But it did not given any clue of ALCAPA, mainly because echo window did not permit visualisation of left coronary artery]. With this background, patient was taken for coronary angiography which was done in multiple views. This revealed typical anatomy of ALCAPA.

DISCUSSION
ALCAPA or Blannd-Garland-White syndrome is a rare congenital anomaly with incidence of 1 in 3 lac live births, accounting for 0.25% of congenital heart disease. The clinical expression of syndrome results from evolving morphological-functional alterations in pulmonary circulation that occur after birth.1 Soon after birth, resistance of the pulmonary circulation is so high permitting antegrade flow from the pulmonary artery (PA) to left coronary artery (LCA), which perfuses the left ventricle. Therefore, occurrence of sudden death is extremely rare in this age group. As pulmonary vascular

Fig. 1a : Right coronary angiography in RAO position showing filling of left coronary system through collaterals.

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of flow from the ALCAPA into the PA, which constitutes a left-to-right shunt. In older patients, abundant high-velocity flow may be seen in the interventricular septum, as a result of intercoronary collaterals. Two-dimensional echocardiography can equally visualize the actual anatomical origin of the ALCAPA and assess the degree of mitral insufficiency. In the present case, 2-D echo examination revealed trivial mitral regurgitation with left ventricular hypertrophy with preserved systolic and diastolic function and there was no regional wall abnormality. Coronary arteries could not be visualised properly.

Coronary angiography which is considered as the most accurate diagnostic technique was performed since echocardiography could not reveal the abnormality. Three angiographic criteria are established for diagnosing ALCAPA.

They are as follows:
1. Retrograde filling of LCA.
2. Connection of LCA with PA.
3. Absence of LCA originating from aorta.

Since all the three criteria are fulfilled, diagnosis of ALCAPA is confirmed.

The adult form of ALCAPA may occur in children and necessarily occurs in adults in approximately 10-15% of cases. It is characterised by exuberance in collateral coronary circulation, which allows survival until adulthood, with case being reported at the age of 72 years.

Wesselhoeft et al.2 classified the clinical spectrum of ALCAPA as follows:
1. *Infantile Syndrome*: This is the most common form. Patient develops acute episode of respiratory insufficiency, cyanosis, irritability and profuse sweating. Most of them die within two years.
3. *Syndrome of Continuous Murmur*: This occurs in asymptomatic patients with angina pectoris. A continuous murmur results from great volume of blood flowing through collateral branches between right and left coronary arteries.
4. *Sudden Death in Adolescents or Adults*: Most of the patients are asymptomatic, but some may experience angina on exertion, cardiac arrhythmias and sudden death.

Once the diagnosis of ALCAPA is established, early surgical repair for correction of defect and prevention of complications and sequelae inherent in natural history of disease is mandatory.
REFERENCES


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API, Rewa Chapter is organizing 16th MP APICON on 15th October 2006. All members of API and Post-Graduate Students of S.S. Medical College, Rewa are cordially invited to attend the conference.

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